309 Delayed development in newly diagnosed infants with CF: the importance of a broad based physiotherapy program

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Background: Without neonatal screening programs for CF, infants with severe respiratory involvement, FTT due to malabsorption and delayed neurological development with hypotonia may be diagnosed late with CF. These infants display a developmental delay until their nutritional and respiratory parameters improve.
Objective: To evaluate if physiotherapy, in addition to standard CF care will improve developmental milestones and respiratory function.
Methods: Chest physiotherapy and neuro-developmental therapy according to Bobath's method, evaluation by AIMS scoring, high fat high caloric diets, intravenous and inhaled antibiotic therapies
Results: Two infants, ages 18 months and 7 months respectively, with CF with severe FTT and severe lung disease were treated. Both suffered from severe hypotonia, developmental delay and were dependent on supplemental oxygen for an extended period resulting in difficulties reaching developmental milestones. Under treatment, the 1st child showed impressive progress at all levels, including decreased oxygen support and weight gain. He could not sit by himself; crawl or transfer between positions (neurodevelopment score (NDS) of a 5–6 months old infant). At age 3.5 years he walks by himself on a wide base, transfers between positions with hands support, plays and talks almost normally for his age (NDS of 2–3 year toddler). The developmental progress of the 2nd child was slow. He died unexpectedly at home at age 15 months.
Conclusions: A broad based physiotherapy program will substantially improve the reaching of developmental milestones in addition to the respiratory condition. Greater attention must be given to the possibility of developmental delay in infants with severe FTT who are diagnosed late with CF.

310* A proactive approach pays off in screened infants with CF

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Background: CF patients demonstrate lung inflammation early in life (Harm 2006). In 2003 neonatal screening for CF was introduced in Scotland. This programme ensures early diagnosis of CF. At RHSC Glasgow we advocate early intervention with respiratory care to try to delay the onset of irreversible lung damage. Our current physiotherapy modalities of choice are Positive Expiratory Pressure therapy (PEP) and Assisted Autogenic Drainage (AAD). We abandoned Postural Drainage and Percussion (PD/Perc) in 2005 as our primary aim is to ensure adequate airflow through all generations of airways, preventing micro-atelectasis. We report results of a retrospective audit of respiratory exacerbation admission rates within the first year of life in CF infants diagnosed by neonatal screening comparing the modalities PD/Perc and PEP/AAD.
Methods: Since February 2003 there have been 32 infants diagnosed with CF in our Region through neonatal screening. For the purpose of this audit we excluded those who were complicated by abdominal surgery due to meconium ileus.
Prior to the change in physiotherapy modality 18 patients received daily PD/Perc. From 2005 a further 14 patients received daily PEP/AAD. Admission rates for respiratory exacerbation were observed in both groups.
Results: The total number of admissions for respiratory exacerbations in the PD/Perc group was 30 compared to the PEP/AAD group where the total number of admissions was 10. The mean number of admissions for the group receiving PD/Perc was 1.667 (SD 2.029). The mean number of admissions with PEP/AAD was 0.71 (SD 1.326).
Discussion: Our results suggest that changing the physiotherapy approach had a positive influence on the number of admissions for exacerbation during the first year of life in screened CF infants.

311 Current practice in prescription, assessment and use of oxygen therapy in cystic fibrosis – a national UK survey

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Background and Aims: Oxygen supplementation therapy has been a standard of care for CF individuals with advanced lung disease despite the current little information on its use and prescription. The aim of this survey is to examine the current practices in UK CF units for prescribing oxygen therapy.
Methods: We sent a questionnaire to lead respiratory/CF consultants caring for CF patients in the UK. Items asked were, indications, prescribing guidelines and current practices.
Results: Of the 160 questionnaires sent, 84 replies were received, 68% were from paediatric units, 26% adult and 6% cared for both age groups (accounting for approximately 4463 CF patients).
As anticipated, adult centres provided oxygen to a greater proportion of their patients compared to paediatric units (5.4% and 1.9% respectively). The commonest indications were hypoxia, breathlessness and terminal care.
The commonest outcome measures used for prescribing Long term oxygen therapy (LTOT) were overnight oximetry and oxygen saturation. Educational support and psychological assessment was considered by 69% and 56% of respondents' units respectively.
Almost all units were not influenced by cost of prescribing any form of Oxygen therapy. 90% of respondent units did not believe that LTOT prolonged survival. The commonest indications for prescribing Short burst oxygen therapy (SBO) and Ambulatory oxygen (AO) were decreased exercise tolerance due to breathlessness (70%) and hypoxia (61%).
Conclusions: Many CF units in the UK provide Oxygen therapy according to clinical need and appear to be un-inhibited by cost. The units show some common practices and themes surrounding oxygen usage among CF patients although there still appears a lack of consensus, highlighting a need for further research and standards.

312 An audit of the importance of nose clips for spirometry in CF

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Background: Although ATS guidelines recommend wearing nose clips during spirometry, currently not all of our patients are agreeable to using nose clips during spirometry.
Aim: To assess if there was a difference in spirometry performed with and without nose clips.
Methods: 10 adults with CF from the Regional Adult CF Centre performed two sets of spirometry 20 minutes apart. Each set consisted of three forced expiratory maneuvers, one with and one without nose clips in a randomised order.
Data Analysis: The mean difference (MD), standard deviation (SD) and limits of agreement were measured to assess the level of agreement between spirometry results with and without nose clips.
Results: 10 subjects (9F:1M), with a mean (SD) age of 21.4 (4.89) yrs, completed the study. There appears to be no significant difference in spirometry results performed with nose clips compared to spirometry performed without nose clips (Table 1).
Conclusion: Although these results need to be verified in a much larger longitudinal study this preliminary data suggests that not wearing nose clips does not affect spirometry results.

Table 1

	Difference, with nose-clips – without nose-clips		
	Mean	SD	Limits of agreement
FEV_1 (L)	0.01	0.14	0.28 to 0.29
FEV_1 (%)	0.20	4.21	−8.22 to 8.62
FVC (L)	0.64	0.25	0.14 to 1.14
FVC (%)	1.50	6.20	−10.91 to 13.91
PEF (L)	−7.30	27.17	−61.65 to 47.05
PEF (%)	−1.20	6.11	−13.41 to 11.01
FEF (L)	−0.13	0.42	−0.98 to 0.72
FEF (%)	−3.00	10.40	−23.81 to 17.81